Agency for Healthcare Research and Quality
Advancing Excellence in Health Care

NATIONAL QUALITY MEASURES CLEARINGHOUSE

General

Title

Amyotrophic lateral sclerosis (ALS): percentage of patients diagnosed with ALS who are dysarthric who were offered a referral at least once annually to a speech language pathologist for an augmentative/alternative communication evaluation.

Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

Measure Domain

Primary Measure Domain

Clinical Quality Measures: Process

Secondary Measure Domain

Does not apply to this measure

Brief Abstract

Description

This measure is used to assess the percentage of patients diagnosed with amyotrophic lateral sclerosis (ALS) who are dysarthric who were offered a referral at least once annually to a speech language pathologist for an augmentative/alternative communication evaluation.

Rationale

Communication is vital to quality of life and most amyotrophic lateral sclerosis (ALS) patients lose the ability to communicate (Ball, Beukelman, & Pattee, "Communication," 2004; Ball, Beukelman, & Pattee, "Acceptance," 2004). Both high tech and low tech options are available through a speech language pathologist to enhance continued communication (Beukelman, Fager, & Nordness, 2011). Dysarthria is present in nearly all ALS patients with bulbar onset and in nearly 70% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-assisted ventilation have a greater need for augmentative alternative communication as the disease progresses (Ball, Beukelman, & Pattee, "Communication," 2004; Ball, Beukelman, & Pattee, "Acceptance," 2004; Beukelman, Fager, & Nordness, 2011).

The following clinical recommendation statements are quoted verbatim from the referenced clinical guidelines and represent the evidence base for

the measure:

- Regular assessment (i.e., every 3 months) of communication by a trained speech therapist is recommended (Andersen et al., 2005).
- The use of appropriate communication support systems (ranging from pointing boards with figures or words, to computerized speech synthesizers) should be provided as required (Andersen et al., 2005).

Evidence for Rationale

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

Andersen PM, Borasio GD, Dengler R, Hardiman O, Kollewe K, Leigh PN, Pradat PF, Silani V, Tomik B, EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral. EFNS task force on management of amyotrophic lateral sclerosis: guidelines for diagnosing and clinical care of patients and relatives. Eur J Neurol. 2005 Dec;12(12):921-38. [122 references] PubMed

Ball LJ, Beukelman DR, Pattee GL. Acceptance of augmentative and alternative communication technology by persons with amyotrophic lateral sclerosis. Augmentative Altern Commun. 2004;20(2):113-22.

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Beukelman D, Fager S, Nordness A. Communication support for people with ALS. Neurol Res Int. 2011;2011:714693. PubMed

Primary Health Components

Amyotrophic lateral sclerosis (ALS); dysarthria; speech language pathology; augmentative/alternative communication evaluation

Denominator Description

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS) who are dysarthric (see the related "Denominator Inclusions/Exclusions" field)

Numerator Description

Patients who were offered a referral at least once annually to a speech language pathologist for an augmentative/alternative communication evaluation

Evidence Supporting the Measure

Type of Evidence Supporting the Criterion of Quality for the Measure

A clinical practice guideline or other peer-reviewed synthesis of the clinical research evidence

A formal consensus procedure, involving experts in relevant clinical, methodological, public health and organizational sciences

A systematic review of the clinical research literature (e.g., Cochrane Review)

One or more research studies published in a National Library of Medicine (NLM) indexed, peer-reviewed journal

Additional Information Supporting Need for the Measure

Importance of Topic

Prevalence and Incidence

- Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig's disease, is a type of motor neuron disease that is a rapidly progressive and fatal neurological disease (National Institute of Neurological Disorders and Stroke [NINDS], 2013).
- Twenty thousand to 30,000 people in the United States (U.S.) have ALS (NINDS, 2013).
- Five thousand people are diagnosed with ALS in the U.S. annually (NINDS, 2013).
- ALS is one of the most common neuromuscular diseases worldwide (NINDS, 2013).
- In 90% to 95% of all ALS cases the disease occurs apparently at random with no clearly associated risk factors (NINDS, 2013).
- Five percent to 10% of all ALS cases are inherited (NINDS, 2013).
- Twenty percent of all familial cases result from a specific genetic defect that leads to mutation of the enzyme known as superoxide dismutase 1 (SOD1) (NINDS, 2013).
- No cure exists for ALS. Newer pharmacotherapy agents have been found to reduce the progression, but not halt the disease development (NINDS, 2013).
- The prevalence of ALS is said to be between six and eight cases per 100,000 in the population. Using the higher prevalence estimate and data from the 2000 U.S. census, nearly 22,600 Americans are living with ALS at any one time. Since ALS is a disease of aging, as the U.S. population increases and ages, an increase in the prevalence of ALS can be anticipated (ALS Association, 2012)
- Cognitive dysfunction is seen in 20% to 50%, while only 3% to 5% develop dementia that is usually of frontotemporal type (Strong et al., 2009). Consensus criteria for diagnosis have recently been reported (Strong et al., 2009).
- Death due to respiratory failure follows on average 2 to 4 years after onset, but a small group may survive for a decade or more (Haverkamp, Appel, & Appel, 1995).
- The mean age of onset is 47 to 52 years in familial cases (FALS) and 58 to 63 years in sporadic (SALS) cases (Bobowick & Brody, 1973).
- The lifetime risk for developing ALS for individuals aged 18 years has been estimated to be 1 in 350 for men and 1 in 420 for women (Armon, 2007) with male sex, increasing age and hereditary disposition being the main risk factors (Heffernan et al., 2006).

Mortality and Morbidity

- Most patients with ALS die within 2 to 5 years of onset (Lechtzin et al., 2002). Only 10% of ALS patients survive for 10 years or more (Miller et al., "Drug, nutritional," 2009).
- Treatment of respiratory insufficiency improves survival, quality of life and respiratory symptoms (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009). The diagnosis and management of respiratory insufficiency is critical because most deaths from ALS are due to respiratory failure (Lechtzin et al., 2002; Miller et al., "Drug, nutritional," 2009; EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis et al., 2012; Laird et al., 2001).
- Falls surveillance will lead to interventions to prevent falls and decrease fall related deaths in ALS patients. Falls are an independent predictor of adverse health outcomes (Gil et al., 2008). Fall related deaths occur in 1.7% of ALS patients (Rubenstein & Josephson, 2002). Several specific risk factors for falls have been identified, including muscle weakness, deficits in gait or balance, visual deficits, arthritis, impairments in activities of daily living, depression, and cognitive impairment (Ringholz et al., 2005).
- Studies confirm the presence of cognitive impairment in 50% of patients with ALS and particularly implicate executive dysfunction and mild memory decline in the disease process (Laird et al., 2001). More severe impairment occurs in a subset of patients with ALS and has features consistent with frontal temporal dementia (FTD) (Phukan, Pender, & Hardiman, 2007; Gordon et al., 2007). Recent studies have demonstrated the feasibility of screening patients in a busy specialized ALS clinic (Flaherty-Craig et al., 2009; Woolley & Katz, 2011), but this is still not routinely practiced. A fuller characterization of the extent of cognitive and behavioral dysfunction in ALS has important implications given that it shortens survival (Elamin et al., 2011), and the burden and stress for carers of patients with FTD is very great. It also has relevance to effective communication, legal issues and end-of-life decision making by patients with motor neuron disease (MND) (Elamin et al., 2011).
- Pseudobulbar affect (PBA), excessive laughing or crying, or involuntary emotional expression disorder affects 20% to 50% of patients with ALS, especially in pseudobulbar palsy (McCullagh et al., 1999). Patients are embarrassed and isolated by these symptoms, which in turn greatly diminishes the patients' quality of life.
- Sialorrhea, or drooling, is embarrassing, socially isolating, and is associated with aspiration pneumonia. The prevalence is estimated at 50%, and 70% of patients receiving oral medications for treatment reported benefit (Laird et al., 2001; Miller et al., "Multi-disciplinary," 2009)
- Fatigue may be a symptom of depression, poor sleep, abnormal muscle activation, immobility, or respiratory dysfunction. Fatigue diminishes quality of life for patients with ALS. Fatigue was a side effect of therapy in 26% of patients taking riluzole vs. 13% taking placebo

(Bensimon, Lacomblez, & Meininger, 1994). Asthenia occurred in 18% of patients taking riluzole vs. 12% of patients taking placebo in a larger study (Lacomblez et al., 1996).

- The prevalence of depression in ALS ranges from 0% to 44%, although systematic studies suggest 10% in advanced ALS (Laird et al., 2001; Wicks et al., 2007). Depression shortens survival and lowers quality of life for patients with ALS (Phukan, Pender, & Hardiman, 2007). There is consensus among experts that depression should be treated in patients with ALS (Laird et al., 2001); however, there are no controlled studies of benefit or harm.
- Insomnia is common in ALS and may be a symptom of early respiratory weakness, underlying anxiety, depression, or pain (Hetta & Jansson, 1997). There is a concern that sedative/hypnotic agents may suppress the respiratory drive in patients with ALS.
- Weight loss is a key prognostic indicator for ALS with the risk of death increased 7-fold when body mass index is less than 18.5 kg/m^2 (Marin et al., 2011; Lehéricey et al., 2012; Spataro et al., 2011; Desport et al., 1999; Vaisman et al., 2009; Dupuis et al., 2008).
- ALS patients have dysarthria in nearly all bulbar onset patients and nearly 40% of ALS patients with spinal onset. More than 95% of ALS patients cannot speak before death and patients who accept gastrostomy tube, non-invasive ventilation or tracheostomy-ventilation have a greater need for augmentative alternative communication as the disease progresses (Ball, Beukelman, & Pattee, "Communication," 2004; Ball, Beukelman, & Pattee, "Acceptance," 2004; Mathy, Yorkston, & Gutmann, 2000; Beukelman, Fager, & Nordness, 2011).
- End of life discussions will improve patient decision making with respect to disease management (NINDS, 2013; ALS Association, 2012; Strong et al., 2009; Haverkamp, Appel, & Appel, 1995; Bobowick & Brody, 1973; Heffernan et al., 2006). Pain in ALS should be treated following accepted guidelines (Oliver et al., 2011; Albert et al., 1999, Mitsumoto et al., 2005; Nolan et al., 2008; Albert et al., 2005; Albert et al., 2009).

Office Visits and Hospital Stays

- One study's significant findings were that common morbidities increased over time (pneumonia [38.1% to 47.3%], respiratory failure [26.9% to 35.5%], and nutritional deficiency [43.0% to 56.3%]); the median length of stay dropped from 6 to 4 days; mean hospital charges increased from $21,574 to $24,314; the proportion of hospital deaths decreased over time (17.6% to 14.6%), whereas the proportion discharged to home health/hospice care (14.0% to 18.2%) and to long-term care facilities (13.2% to 27.9%) increased. The odds ratio (OR) of death was 5.03 (95% CI: 4.57 to 5.54) for those admitted with respiratory failure, 1.36 (1.24 to 1.50) for those with pneumonia, and 0.84 (0.77 to 0.92) for those with nutritional deficiency. The high OR of death in patients admitted for pneumonia or respiratory failure is likely associated with more advanced disease, whereas the protective effect of admission for nutritional deficiency is consistent with the predominance of bulbar symptoms and admission earlier in the disease. The trends during the 15 years of this administrative data set were for increasing comorbidities and higher utilization of end-of-life care (Dubinsky, Chen, & Lai, 2006).

Family Caregiving

- Caregiver burden was correlated to their level of depression and quality of life and, differently from other chronic disorders, increased with the worsening of patients' disability. ALS patients have a good objective perception of their impact on caregivers (Chiò et al., 2005).
- Recent studies assessing caregivers' burden in chronic neurologic disorders have found some features shared by caregivers: the perceived burden exceeds the objective measures of patients' impairment, the amount of burden is independent of diagnosis, and the patients' cognitive functioning is an important factor in determining the level of burden (Thommessen et al., 2002).

Cost

- ALS is a difficult to diagnose, fatal, progressive degenerative disease with an average survival time of 2 to 5 years. Percutaneous endoscopic gastrostomy (PEG) and bi-level intermittent positive pressure (BIPAP) ventilation may be the major interventions leading to longer survival of patients with ALS. Riluzole has been shown to have modest effects on survival (as opposed to functional) gains and is currently the only drug approved for the treatment of ALS. Mechanical ventilation (via a tracheostomy tube) is expensive, but is widely used in later stage patients with ALS in the U.S. A review of nine cost-effectiveness studies of riluzole found the following: drug costs and survival gains are the major drivers of cost effectiveness; survival gains are estimated from truncated databases with a high degree of uncertainty; more accurate stage-specific utility weights based on patients who agreed to treatment are needed; case incidence-based evaluations should be carried out; cost-effectiveness ratios are insensitive to discount rates; employment and caregiver issues or externalities have been widely ignored; threshold acceptance cost-effectiveness values are ill-defined and evaluations are not generalizable to other countries because of cost and treatment style differences. On account of the high degree of uncertainty pertaining to survival gains and the relatively high costs per life years or quality-adjusted life-years gained, and while acknowledging that not every therapy has to be cost effective (e.g., orphan drugs), it is still inconclusive as to whether or not riluzole can be considered as cost-effective therapy for ALS (Ginsberg & Lowe, 2002).

Disparities

- All races and ethnic backgrounds are affected by ALS (NINDS, 2013).

- ALS most common in individuals 40 to 60 years old, but younger and older people can develop the disease (NINDS, 2013).
- Men are more likely to develop ALS than women. Studies suggest an overall ratio of about 1.5 men to every woman who develops ALS in Western countries (ALS Association, 2012).

Opportunity for Improvement

- Speech assessment in ALS patients identifies dysarthria, independent of dysphagia, that limits communication and maintenance of their active communicator role. Speech correction should focus on the maintenance of functional communication. Assessment should involve measurement of speech rate (words per minute). Augmentative and alternative communication is recommended when speech rate is less than 125 words per minute. Nearly 88% of ALS patients are evaluated by this criterion, but fewer than half implement appropriate interventions (Lévêque, 2006; Robert et al., 2006; American Speech-Language-Hearing Association [ASHA], 2002; Beukelman, Fager, & Nordness, 2011; Hanson, Yorkston, & Britton, 2011).

Evidence for Additional Information Supporting Need for the Measure

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Extent of Measure Testing

This measure is being made available without any prior testing. The American Academy of Neurology (AAN) recognizes the importance of testing of all of its measures and encourages testing of the amyotrophic lateral sclerosis (ALS) measurement set for feasibility and reliability by organizations or individuals positioned to do so. The AAN welcomes the opportunity to promote the initial testing of these measures and to ensure that any results available from testing are used to refine the measures before implementation.

Evidence for Extent of Measure Testing

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

State of Use of the Measure

State of Use

Current routine use

Current Use

not defined yet

Application of the Measure in its Current Use

Measurement Setting

Measurement Setting

Ambulatory/Office-based Care

Home Care

Hospital Outpatient

Skilled Nursing Facilities/Nursing Homes

Professionals Involved in Delivery of Health Services

not defined yet

Least Aggregated Level of Services Delivery Addressed

Individual Clinicians or Public Health Professionals

Statement of Acceptable Minimum Sample Size

Does not apply to this measure

Target Population Age

Unspecified

Target Population Gender

Either male or female

National Strategy for Quality Improvement in Health Care

National Quality Strategy Aim

Better Care

National Quality Strategy Priority

Person- and Family-centered Care
Prevention and Treatment of Leading Causes of Mortality

Institute of Medicine (IOM) National Health Care Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness

Patient-centeredness

Data Collection for the Measure

Case Finding Period

Unspecified

Denominator Sampling Frame

Patients associated with provider

Denominator (Index) Event or Characteristic

Clinical Condition

Denominator Time Window

not defined yet

Denominator Inclusions/Exclusions

Inclusions

All patients with a diagnosis of amyotrophic lateral sclerosis (ALS) who are dysarthric

Note: Refer to the original measure documentation for International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) diagnosis codes and Current Procedural Terminology (CPT) Evaluation and Management (E/M) service codes.

Exclusions

Documentation of a medical reason for not offering a referral to a speech language pathologist for an augmentative/alternative communication evaluation (e.g., patient is already using an augmentative communication device).

Exclusions/Exceptions

not defined yet

Numerator Inclusions/Exclusions

Inclusions

Patients who were offered a referral at least once annually to a speech language pathologist for an augmentative/alternative communication evaluation

Exclusions

Unspecified

Numerator Search Strategy

Fixed time period or point in time

Data Source

Administrative clinical data

Electronic health/medical record

Paper medical record

Type of Health State

Does not apply to this measure

Instruments Used and/or Associated with the Measure

Unspecified

Computation of the Measure

Measure Specifies Disaggregation

Does not apply to this measure

Scoring

Rate/Proportion

Interpretation of Score

Desired value is a higher score

Allowance for Patient or Population Factors

not defined yet

Standard of Comparison

not defined yet

Identifying Information

Original Title

Measure #9: ALS communication support referral.

Measure Collection Name

Amyotrophic Lateral Sclerosis Performance Measurement Set

Submitter

American Academy of Neurology - Medical Specialty Society

Developer

American Academy of Neurology - Medical Specialty Society

Funding Source(s)

Unspecified

Composition of the Group that Developed the Measure

Work Group Members Amyotrophic Lateral Sclerosis

Co-Chairs: Robert G. Miller, MD; Benjamin Rix Brooks, MD

American Academy of Neurology Representatives: Steven Ringel, MD; Hiroshi Mitsumoto, MD; Carlayne Jackson, MD; Christen Shoesmith, MD, BSc; Edward Kasarskis, MD, PhD

Pulmonologist: Robert C. Basner, MD

Gastroenterologist: Nicholas Procaccini, MD

American Academy of Physical Medicine and Rehabilitation: Gregory Carter, MD

Nurse: Dallas Forshew, RN, BSN

Physical Therapy/Occupational Therapy: Mohammed Sanjak, PhD, PT, MBA; Pat Casey, MS, CRCC

Hospice/Palliative Care Specialist: Bob Osborne, RN

Muscular Dystrophy Association: Valerie Cwik, MD

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Financial Disclosures/Other Potential Conflicts of Interest

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Adaptation

This measure was not adapted from another source.

Date of Most Current Version in NQMC

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Measure Maintenance

Unspecified

Date of Next Anticipated Revision

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Measure Status

This is the current release of the measure.

Measure Availability

Source available from the American Academy of Neurology (AAN) Web site .

For more information, contact AAN at 201 Chicago Avenue, Minneapolis, MN 55415; Phone: 800-879-1960; Fax: 612-454-2746; Web site: www.aan.com .

NQMC Status

This NQMC summary was completed by ECRI Institute on March 8, 2016. The information was not verified by the measure developer.

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Production

Source(s)

American Academy of Neurology (AAN). Amyotrophic lateral sclerosis performance measurement set. St. Paul (MN): American Academy of Neurology (AAN); 2012 Jul 31. 75 p.

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